Operative Techniques in
Thoracic and
Cardiovascular Surgery

Tetralogy of Fallot with Atrioventricular Canal Defect: One Patch Repair

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The incidence of tetralogy of Fallot (TOF) among patients with atrioventricular septal defect (AVSD) is estimated to be about 6% to 10%.[1,2] This combination of lesions poses certain unique structural and physiological challenges to the surgeon. Opinions differ regarding the timing of surgery—including the need for a palliative systemic-to-pulmonary artery shunt prior to complete repair, the use of a single versus multiple patches to address the septal defects, the surgical approach to relieve right ventricular outflow tract (RVOT) obstruction, and the effects of performing a transannular patch (TAP).

Studies mention a high incidence of trisomy 21 (60%-90%)[1-7] in this subgroup. Rastelli type C is the predominant type of AVSD in this association.[4,6,8] Also, the incidence of left ventricular outflow tract (LVOT) obstruction seems lower than in AVSD without associated TOF due to the anterior displacement of the conal septum. The presence of RVOT obstruction seems to protect the patient from the pulmonary congestion often seen in AVSD. Historically, a systemic-to-pulmonary artery shunt has often been performed as the first procedure to palliate the cyanosis associated with this condition.

Although operative techniques differ, early morbidity usually stems from residual shunts and cardiogenic shock.[2,4-6] Freedom from late reoperation averages about 65% to 80% and repair/replacement of the left atrioventricular valve (LAVV) for regurgitation is the most commonly performed procedure.[4,5,7] Surgery for late LVOT obstruction is unusual.[3,4,7]

Free pulmonary valve insufficiency after a TAP causing volume overload of the right ventricle is a concern raised by many authors. This, in concert with significant right atrioventricular valve regurgitation and an infundibulotomy, may be a source of strain for the right ventricle. Hence, some argue for a valve-sparing transatrial-transpulmonary approach.[5] Others, however, have not found any significant mid- or long-term adverse effects from an infundibulotomy or a TAP.[1,7]

Our approach has been unchanged since the publication of the group's early experience in 1998.[8] We prefer a complete correction in early infancy, between 3 and 6 months of age. We do not believe that a systemic-to-pulmonary artery shunt is required for palliation. We use a single patch to repair the septal defects and routinely divide the superior bridging leaflet to visualize the ventricular septal defect (VSD) better. The division is performed just to the right of the midline to follow the crest of the malaligned ventricular septum, as described by Vargas et al, to prevent LVOT narrowing.[9] The LAVV cleft is routinely closed.

We perform a transatrial RVOT myectomy and employ a limited infundibulotomy, if necessary. Pulmonary valvotomy, infundibular patch, or a transannular patch with or without a monocusp valve or a right ventricle to pulmonary artery homograft conduit is used as deemed necessary by the surgeon.

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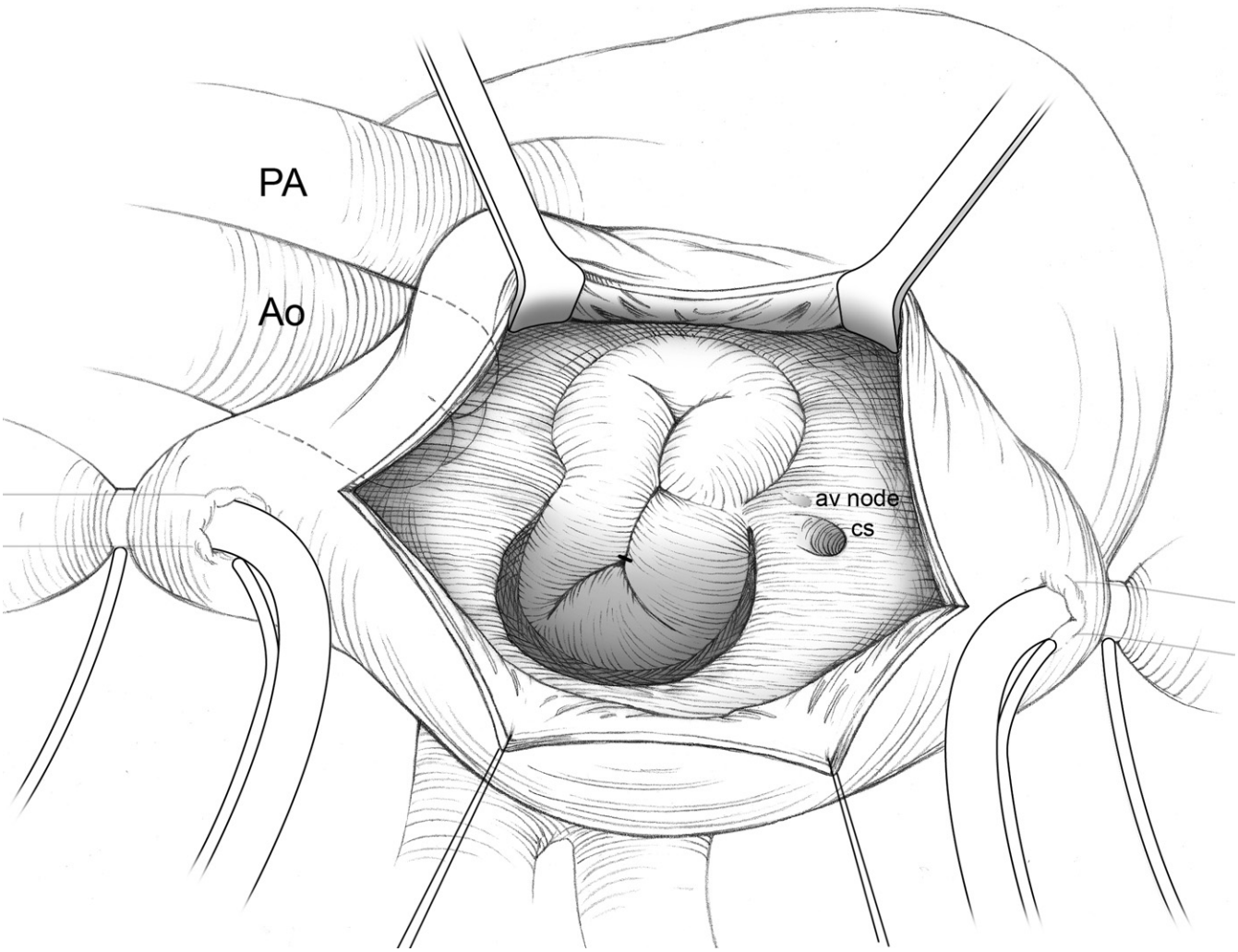

Figure 1 A standard median sternotomy is performed and moderate hypothermic (25°C) cardiopulmonary bypass is instituted with aortic and bicaval cannulation. A large piece of autologous pericardium is harvested and tanned in gluteraldehyde solution. Cold crystalloid cardioplegia is used and a left ventricular vent is placed via the right superior pulmonary vein. An oblique right atriotomy is performed and the anatomy is visualized. If there is any difficulty inspecting the LAVV, the bridging tissue between the ostium primum and the patent foramen ovale/ostium secundum is divided, with care to resuture it with pledgeted sutures prior to the atrial septal defect closure. The atrioventricular (AV) valves are inspected thoroughly with flotation tests to assess competency. The tip of the LAVV cleft is marked with a single 6-0 polypropylene suture. The "tip of the cleft" is defined as the furthest lateral point of insertion of the chordae on the leaflet. Ao = aorta; av = atrioventricular; cs = coronary sinus; PA = pulmonary artery.

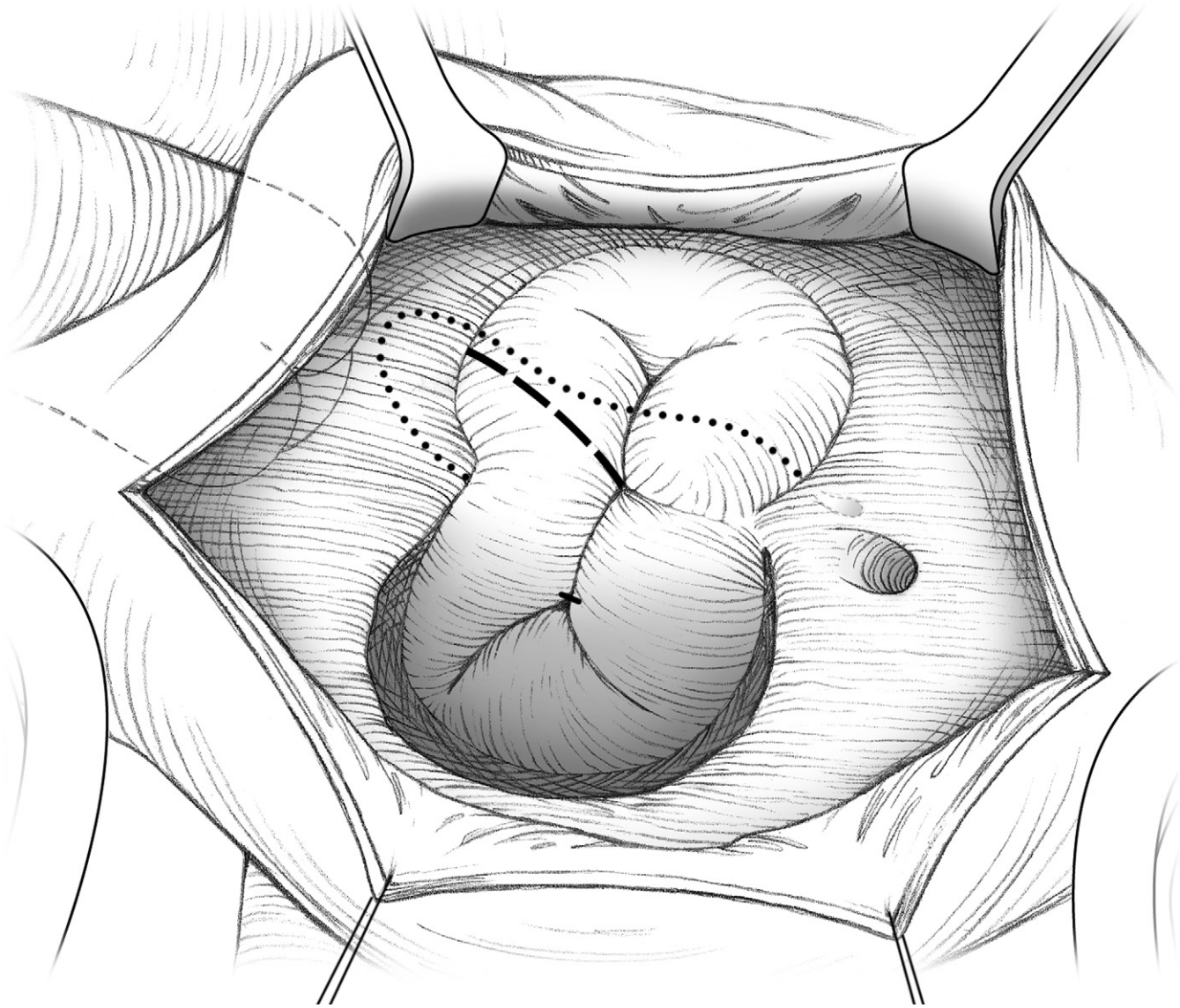

Figure 2 The superior bridging leaflet is then divided from its free edge to the annulus in an oblique fashion, as shown by the dashed line, paralleling the crest of the ventricular septum, shown by the dotted line. In an uncomplicated AVSD, the leaflet division is performed along the midline. However, when there is aortic override and hence is malpositioned to the right, as shown by the dotted line above, angling the leaflet division to the right along the crest of the malaligned septum helps the repair in 2 ways. First, the ventricular component of the patch parallels the natural deviation of the septum better, preventing it from bulging into the LVOT, causing subaortic obstruction. Second, there is less tension on the LAVV component, which would otherwise exist if the division was performed along the middle, with the VSD patch tugging on the bridging leaflets to the deviated septal crest.

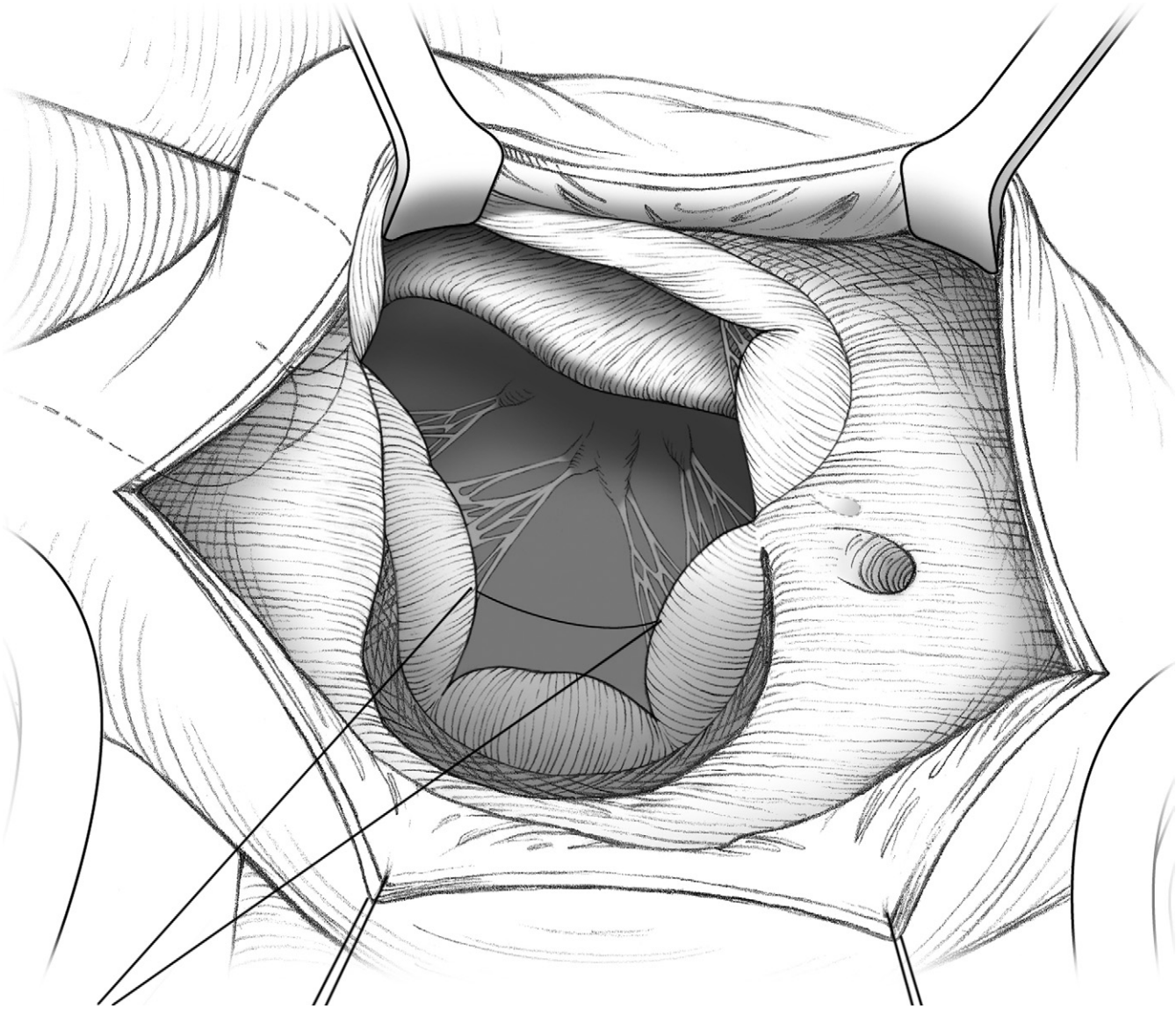

Figure 3 The superior bridging leaflet is almost always Rastelli type C and hence is easily retractable and greatly helps visualization at this stage.

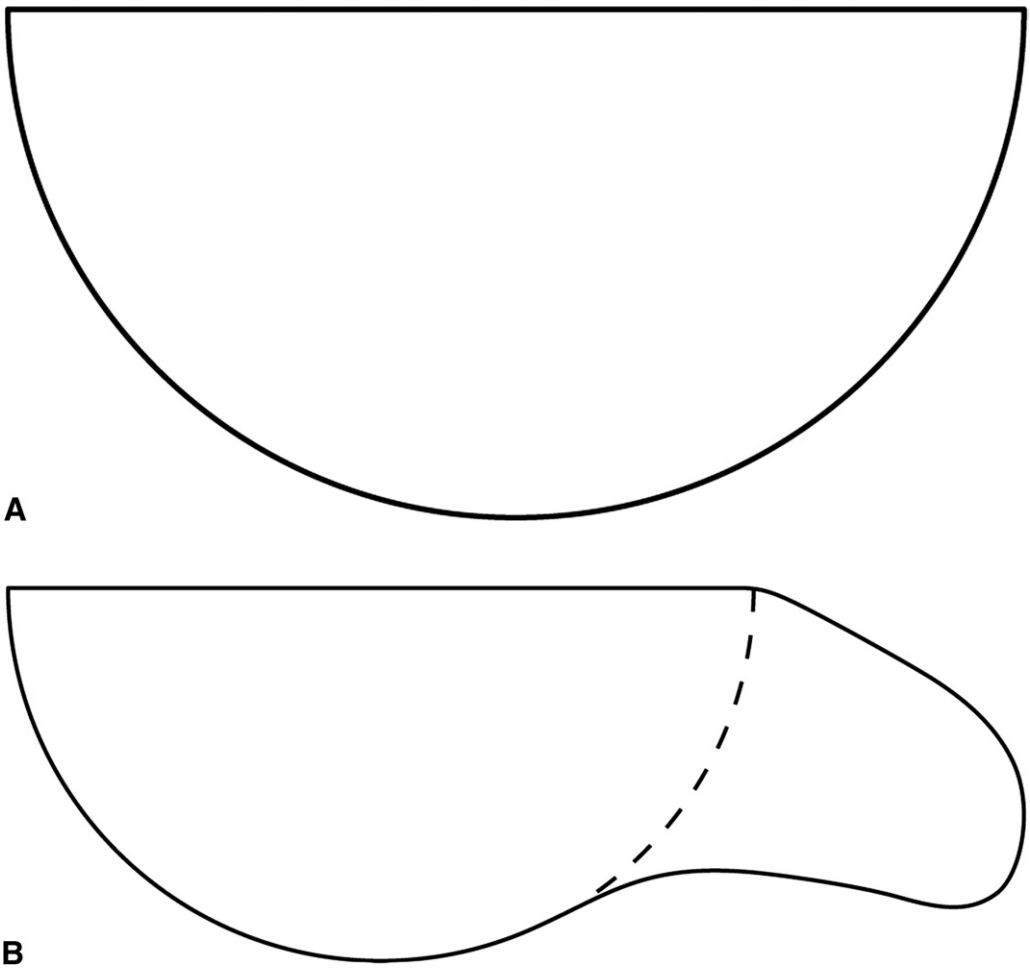

Figure 4 The autologous pericardial patch is sized to the defect with a teardrop-shaped anterior extension. (A) The VSD component of the patch in an otherwise uncomplicated AVSD, and (B) the patch shape in AVSD with TOF. The shape and size of the anterior extension are decided by the size of the VSD and the extent of the aortic override. In general, it is advisable to make the anterior extension a little redundant, oversizing it by about 20% to make more room for the LVOT.

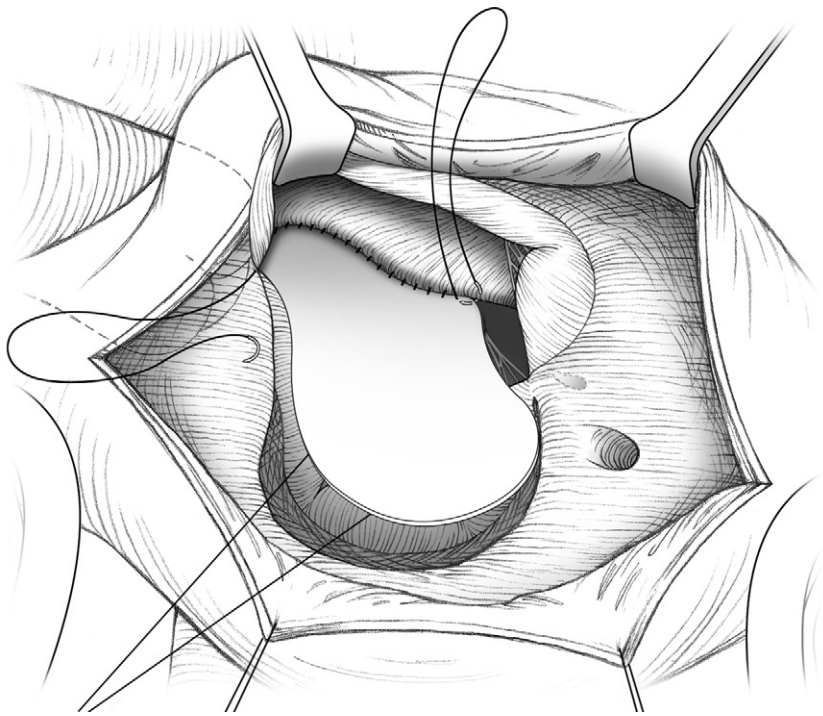

Figure 5 The patch is then sutured in a continuous fashion with 5-0 polypropylene suture taking care to place the bites on the right side of the septum. We start from the most distal point of the defect and suture toward the surgeon with both limbs of the suture. After closing the VSD, the suture ends are placed in nontraumatic clamps. A transatrial infundibulectomy is then performed and hypertrophied RVOT muscle bundles are resected.

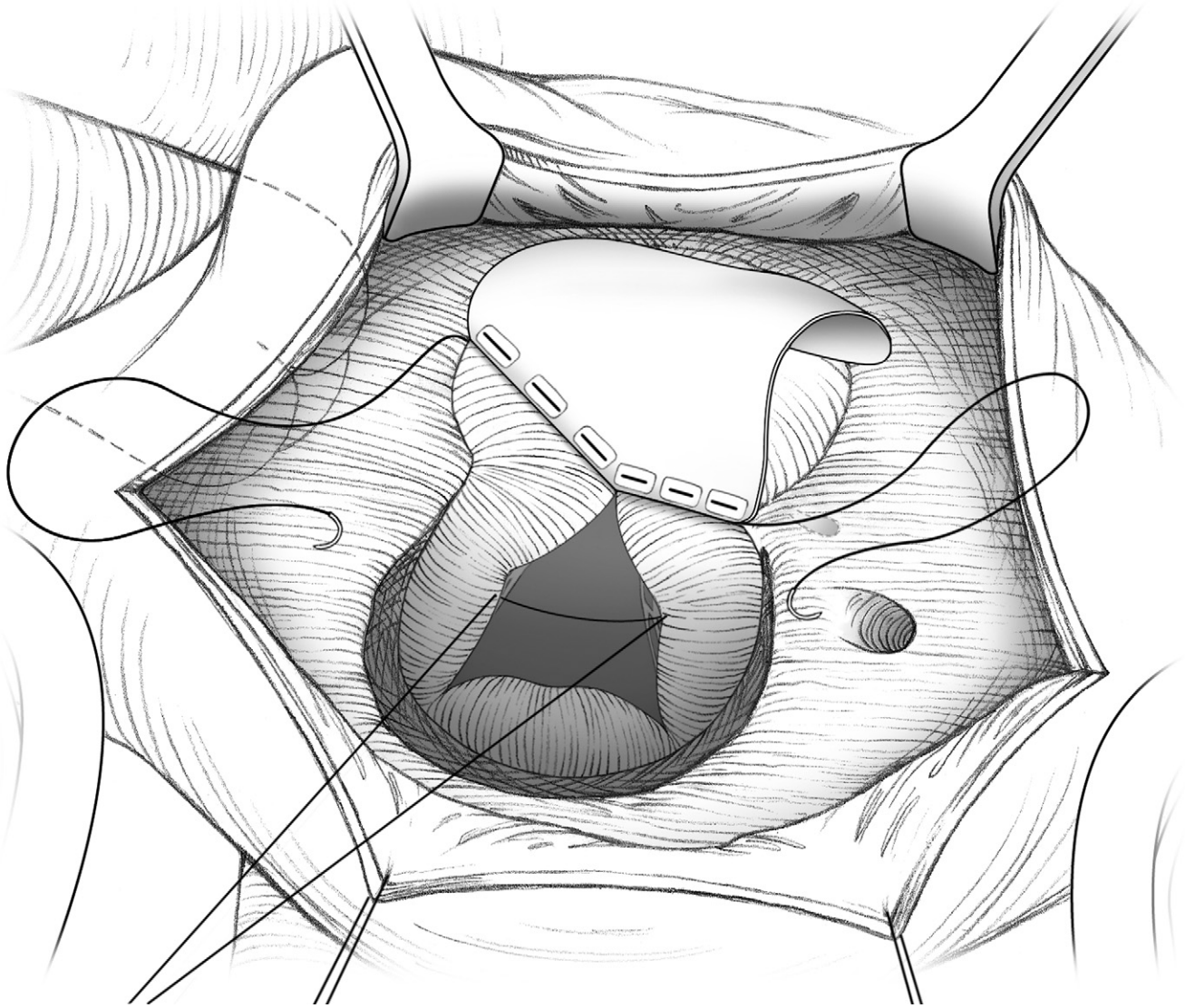

Figure 6 The AV valves are now resuspended to the pericardial patch with multiple interrupted 6-0 polypropylene horizontal mattress sutures. Small autologous pericardial pledgets are used on the left side. The sutures are passed from the left sided leaflet, through the patch and then through the right sided leaflet with knots on the right atrial aspect. The leaflet incision having been placed more on the rightward aspect of the SBL spares more of the leaflet on the left side for a comfortable repair.

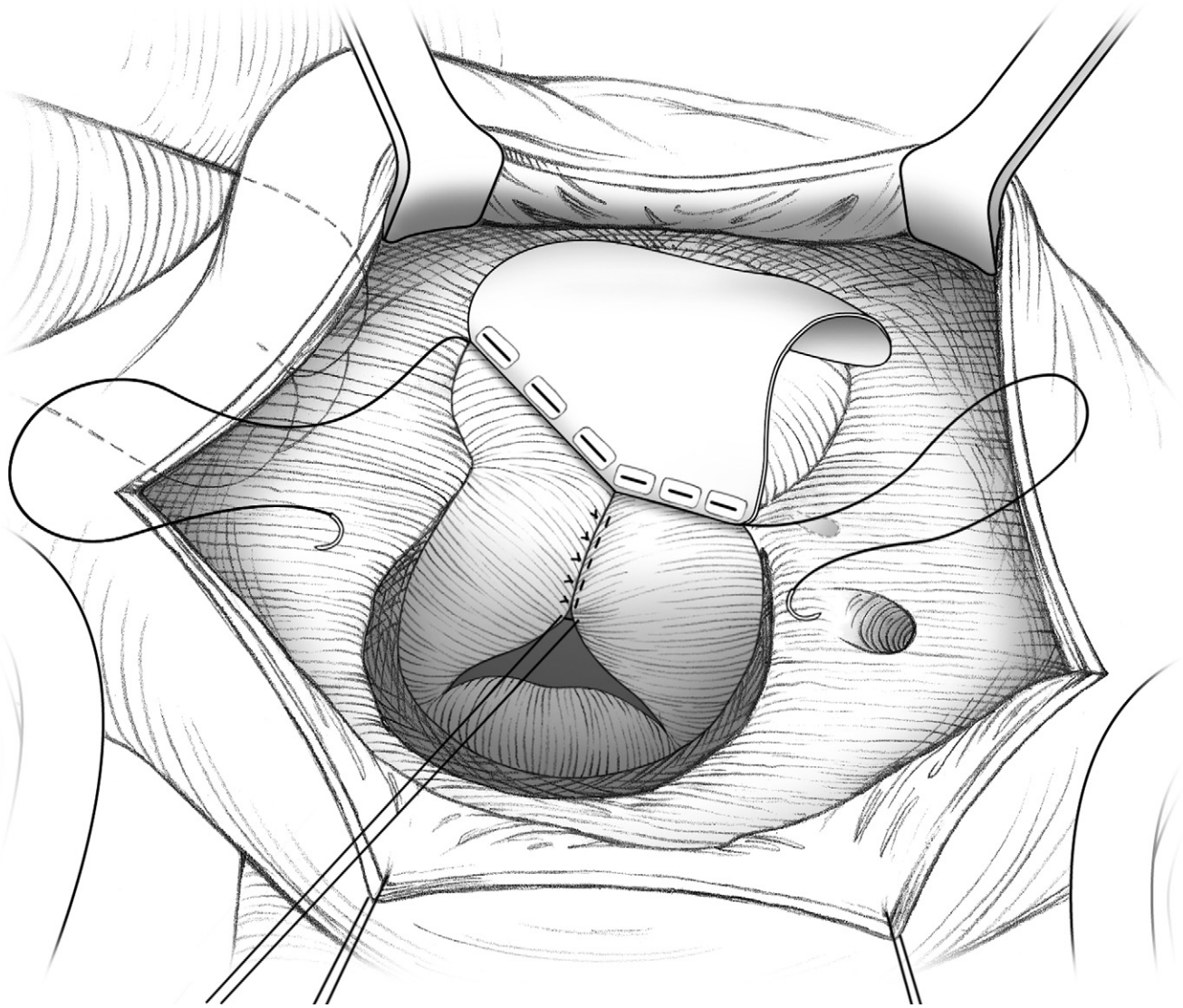

Figure 7 The cleft in the left AV valve is then closed with multiple interrupted 6-0 polypropylene sutures and tested again for competency. Reed annuloplasty or commissuroplasty is performed as deemed necessary. Dilators are used to size the valve orifice to the appropriate *Z*-score if there is any concern for stenosis.

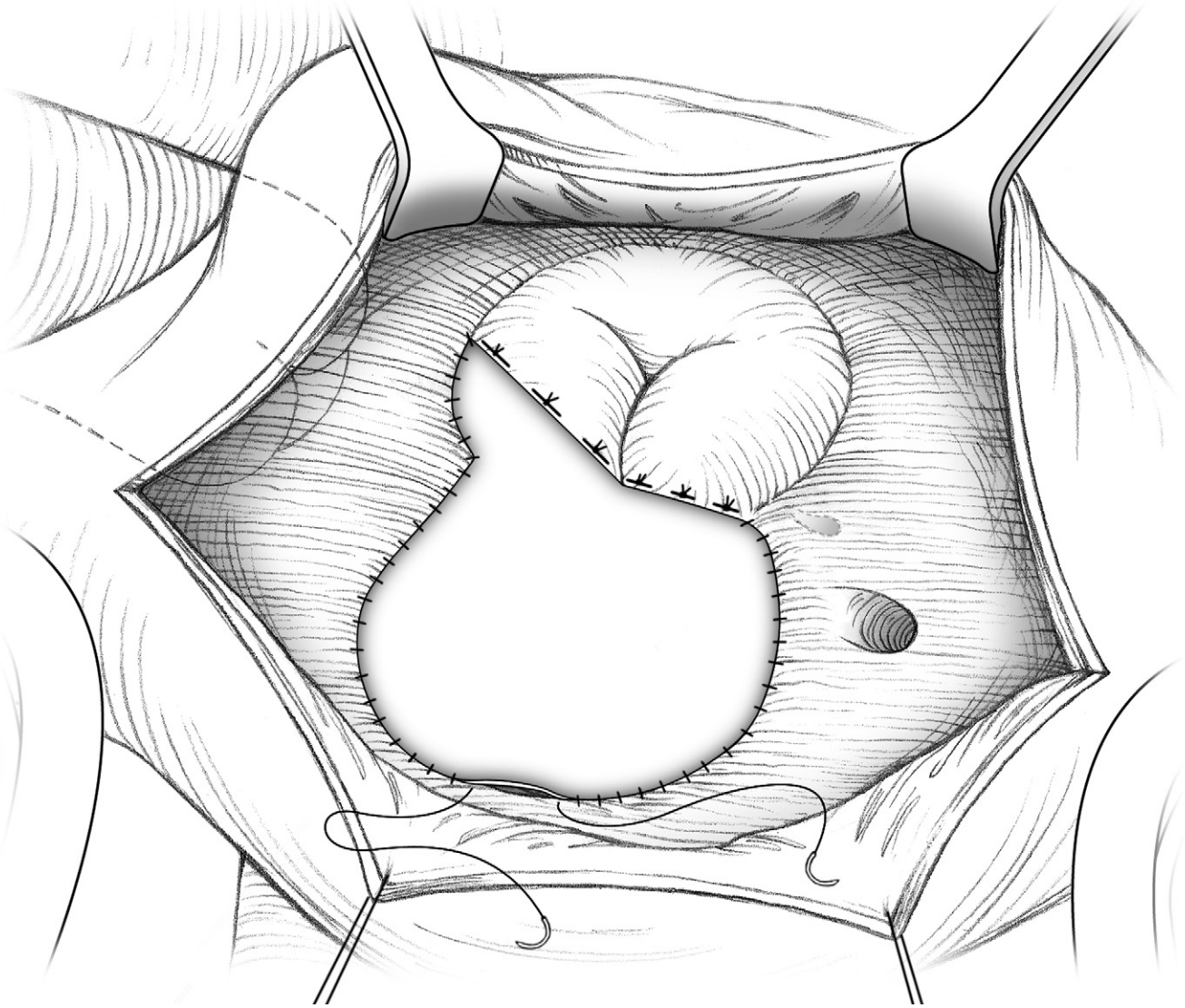

Figure 8 The pericardial patch is then sutured to the edge of the atrial septal defect (ASD) leaving the coronary sinus ostium on the right atrial aspect. The right AV valve is now leak-tested and repaired with commissuuroplasty sutures if needed. The pulmonary valve is then examined via a longitudinal pulmonary arteriotomy, the annulus sized and a valvotomy performed if needed. The RVOT is addressed either with a limited transannular patch, or with an RV-PA conduit as dictated by the coronary artery pattern. A "cormatrix" (Cormatrix Cardiovascular Inc., Alpharetta, GA) monocusp valve is often fashioned with a TAP. The heart is then deaired, uncrossclamped and the usual steps taken to wean off cardiopulmonary bypass. Intraoperative post-repair transesophageal echocardiography is routinely performed to assess ventricular function, valvar competency and to detect patch leaks.

Conclusions

Early total correction of the combined AVSD and TOF lesions can be performed with a low mortality and morbidity, especially in the current era, as observed by Prifti et al.[10] The traditional concerns of postrepair right ventricle dysfunction from volume overload have not been observed consistently. The single-patch technique mentioned above can be used successfully in most circumstances. However, in children less than 3 to 4 months of age or in patients without trisomy 21, there is often not enough redundant atrioventricular valve tissue, necessitating a 2-patch repair. Also, in patients where the override is severe and closer to double outlet right ventricle, we prefer a 2-patch technique to avoid compromising the tricuspid valve. As mentioned earlier, the most common reason for late reoperation has been LAVV regurgitation and not LVOT obstruction.

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